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Clinical Study

Transcatheter closure as an alternative to surgical management in six cases of residual intramural ventricular septal defects after repair of conotruncal malformations

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KEYWORDS

Conotruncal anomaly;
Device closure;
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Abstract *Objectives:* The aim of this study was to determine whether transcatheter closure could be used as an alternative to surgical management in intramural ventricular septal defect, a rare complication after surgical repair of a conotruncal anomaly.

Methods: We reviewed the echocardiographs, catheterization reports and clinical files of children with one or more residual defects of the ventricular septum after repair of conotruncal anomalies seen at the Prince Sultan Cardiac Centre, Riyadh, Saudi Arabia. Between 2 years, six children with residual intramural shunts were identified. Three cases occurred after repair of the tetralogy of Fallot and three after repair of a double outlet in the right ventricle.

Results: Two of the patients underwent surgical closure of the ventricular septal defect, two underwent device closure by catheterization, and two were found to have minimal left-to-right shunt and were managed conservatively. The mean hospital stay was 27.5 days for the surgical patients and 2 days after the transcatheter intervention. No early or late complications were seen in the patients who had device closure, while both surgical patients had prolonged chest drainage, and the post-surgical course of one patient was complicated by dysrhythmia and low cardiac output.

Conclusion: Surgical repair is a successful method of treatment; however, transcatheter closure may offer a less invasive approach. The decision on the management of such patients should be made by the treating physician.

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Introduction

Intramural ventricular septal defect is a well-described clinical entity characterised by intraventricular communication outside the plane of the intraventricular septum. It is a rare post-operative defect after surgical repair of conotruncal anomalies.[1] In conotruncal lesion repairs, incomplete closure of the principal defect, ventricular septal defect patch dehiscence and unrecognized additional defects are the most common causes of post-surgical residual defects. In general, these residual lesions are within the interventricular septum

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plane. Rarely, an unusual type of residual ventricular septal communication is seen in which the interventricular communication lies outside the ventricular septal plane. These defects were first described and named intramural ventricular septal defects in 1994.[1]

Until recently, further cardiac surgery was the only available means of treating intramural ventricular septal defects.[2] In 1999, however, a transcatheter technique for closing such defects was introduced,[3,4] which has subsequently been used widely.[5–7] It is unclear, however, whether the results of a trans-catheter intervention are equivalent to those of surgery. The aim of this study was to determine on the basis of our experience whether transcatheter closure can be used as an alternative to surgery in the treatment of intramural ventricular septal defect.

Case reports

Patient 1

An 8-year-old girl presented with shortness of breath and poor growth, suggesting congestive heart failure. She had no history of fever or constitutional symptoms but had undergone repair of tetralogy of Fallot at the age of 3. On examination, she had tachycardia, hepatomegaly and grade 2/6 systolic murmur in the left lower sternal border. A chest X-ray showed cardiomegaly with increased pulmonary vascular marking. Transthoracic echocardiography showed a residual ventricular septal defect shunt measuring 6 mm in the postero-superior part of the defect patch (Figure 1). Transoesophageal echocardiography before Amplatzer® defect closure showed a 6-mm ventricular intramural septal defect. After device closure, transoesophageal echocardiography showed a good position of the device, with no residual shunt.

Patient 2

A 7-year-old boy presented with difficulty in walking and climbing stairs. The family reported no history of sweating or fever. Double outlet right ventricular repair was performed when the boy was 2 years old. On examination, he had tachypnoea, tachycardia and grade 3/6 systolic murmurs in the left lower sternal border. A chest X-ray showed mild cardiomegaly and mildly increased pulmonary vascular markings. Transthoracic echocardiography showed two residual shunts: a 5-mm ventricular septal defect shunt in the postero-superior part of the defect patch and an additional left ventricle to right atrium shunt, known as a Gerbodi defect. Transoesophageal echocardiography by catheterization before Amplatzer® closure confirmed the 5-mm intramural ventricular septal defect. After device closure, transthoracic echocardiography (Figure 2) showed a good device position with no residual shunt. The Gerbodi defect was considered by the interventionist and the treating physician not to be haemodynamically significant.

Patient 3

A 9-year-old boy presented with dyspnoea on exertion. He had undergone repair of tetralogy of Fallot at the age of 4. Physical examination and a chest X-ray gave findings similar to those for patient 2. Transthoracic echocardiography showed a residual (6–7 mm) intramural ventricular septal defect shunt in the postero-superior defect patch. Pre-operative transoesophageal echocardiography in the operating room before surgical defect closure confirmed the transthoracic findings. After surgical

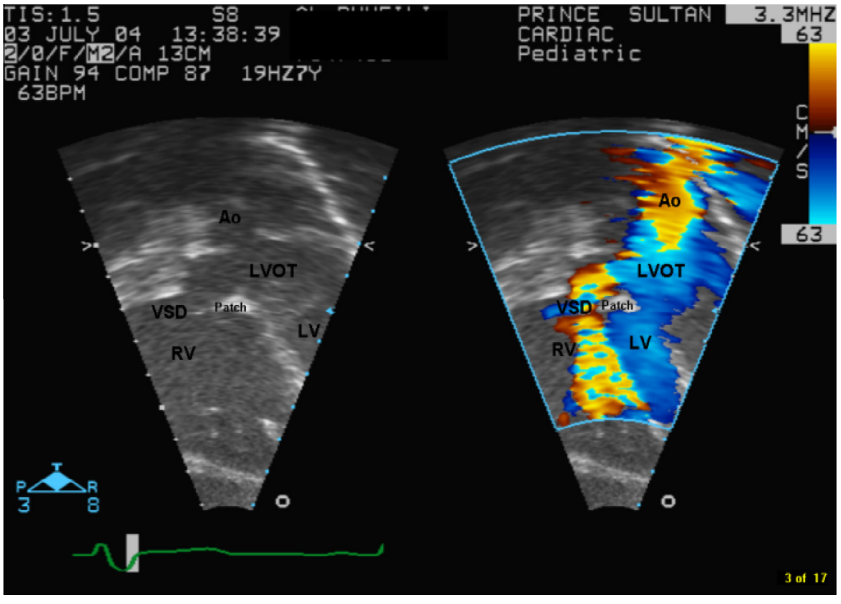

Figure 1: Pre-ventricular septal defect device deployment. Colour Doppler used to compare flow demonstrates residual interventricular communication between the upper margin of the ventricular septal defect patch and the free right ventricle wall. Ao, aorta; LV, left ventricle; LVOT, left ventricular outflow tract; RV, right ventricle; VSD, ventricular septal defect.

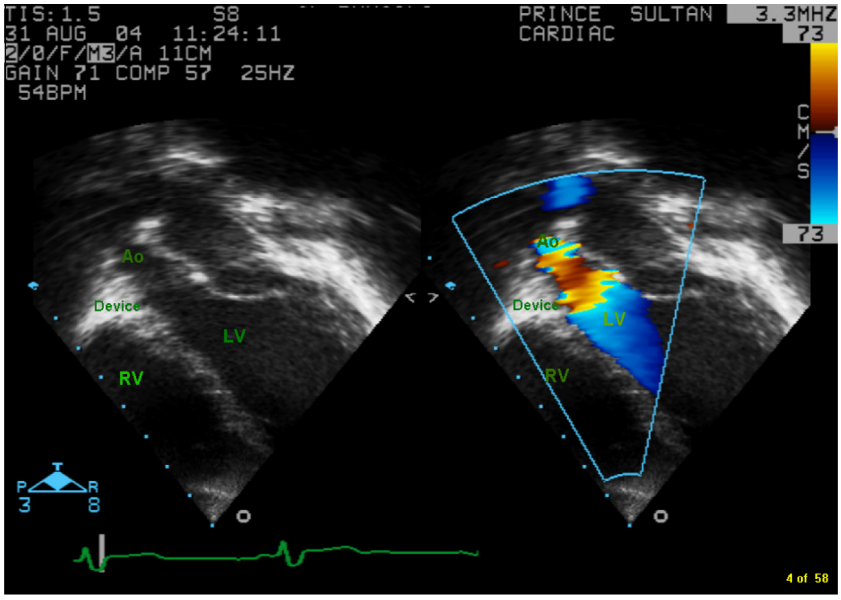

Figure 2: Post-ventricular septal defect device closure. The position of the device appears to be stable. Colour flow mapping confirmed the absence of residual shunting through the inter-trabecular channels between the upper margin of the ventricular septal defect patch and the free RV wall. Ao, aorta; LV, left ventricle; RV, right ventricle.

closure, transoesophageal echocardiography showed no residual shunt across the new patch.

Patient 4

A 7-year-old girl presented with dyspnoea on exertion and palpitations. She had undergone repair of a double outlet right ventricular defect and malposed great arteries at the age of 3. The findings of the physical examination and chest X-ray were similar to those of patients 2 and 3. Transthoracic echocardiography showed a residual (5-mm) intramural ventricular septal defect in the postero-superior defect patch, which was confirmed by pre-operative transoesophageal echocardiography. After surgical defect closure, there was no residual shunt on transoesophageal echocardiography.

Both patients 1 and 2, who underwent transcatheter device closure, and patients 3 and 4, who underwent surgical intervention, were examined by transoesophageal echocardiography and right and left heart catheterisation. The significance of the residual ventricular septal defect was assessed by colour Doppler flow mapping and quantitative shunt calculation. Multiple angiographic projections were performed to profile the defect (Figure 3A) from their geometry and relation to the surrounding structure. The indications for closure were signs and symptoms of heart failure and/or a significant pulmonary-to-systemic flow ratio (Qp:Qs $\geqslant$ 2.0), estimated by echocardiography or at cardiac catheterisation.

Patients 1 and 2 underwent device closure with Amplatzer® muscular devices (Figure 3B). The technique used was that described previously.[4] Patients 3 and 4 underwent surgical closure of their septal defect. The decision to perform surgical closure or device closure was made by the treating physician. The mean hospital stay was 27.5 days for the surgical patients and 2 days for the transcatheter intervention. No early or late complications were seen in the patients who had device clo-

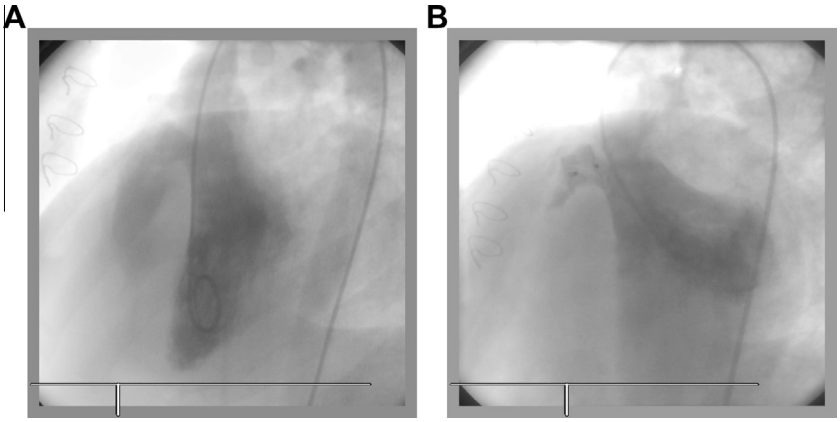

Figure 3: (A) Long axial view of still-frame angiogram of the left ventricular outflow tract with the intramural ventricular septal defect. (B) As in A after Amplatzer® muscular ventricular septal occluder device closure, with minimal residual shunt seen through the device.

Table 1: Demographic and clinical data for the six patients.

Patient No.	Sex	Age (years)	Diagnosis	Age at first operation (years)	Age at diagnosis of defect (years)	Treatment offered
1	F	8	Tetralogy of Fallot	3	5	Device
2	M	7	Double-outlet right ventricle, sub-aortic ventricular septal defect	2	4	Device
3	M	9	Tetralogy of Fallot	4	6	Surgical
4	F	7	Double-outlet right ventricle, malposed great artery, and severe sub-pulmonic stenosis	3	4	Surgical
5	M	5	Tetralogy of Fallot, right ventricular outflow obstruction	1.5	2	Conservative
6	F	7	Double-outlet right ventricle	2.5	4	Conservative

sure. The post-surgical course of patient 3 was complicated by dysrhythmia and low cardiac output, and both surgical patients had prolonged chest drainage.

All four patients were followed-up regularly for 3 years. They showed complete clinical and echocardiographic resolution, with no residual intramural ventricular septal defect shunt. At the last outpatient visit, there was no evidence of arrhythmia or heart block in either the surgical or the device group. No major residual defects were detected at the last echocardiography. All four patients were asymptomatic and were not on regular medication.

Patient 5

A 5-year-old boy had undergone repair of the tetralogy of Fallot with significant right ventricular outflow obstruction at the age of 1.5 years. He was asymptomatic, with no signs of congestive heart failure clinically, but was found incidentally by transthoracic echocardiography during routine outpatient follow-up to have a 3-mm intramural ventricular septal defect.

Patient 6

A 7-year-old girl had undergone double outlet right ventricular repair at the age of 2.5 years. Like patient 5, she was asymptomatic, with no signs of congestive heart failure clinically, but was found incidentally by transthoracic echocardiography during routine outpatient follow-up to have a 4-mm intramural ventricular septal defect.

The chest X-rays of both patients were normal, and the treating cardiologist and his team decided to follow them conservatively. They were followed-up regularly for 3 years and were found to be clinically stable, with no history or clinical examination suggestive of congestive heart failure. Electrocardiograms were unremarkable for all patients at their last clinic visit.

The demographics and clinical data for the six children with intramural ventricular septal defects are listed in Table 1.

Discussion

In this review of six cases we found considerable morbidity in the two surgically treated patients, while transcatheter device closure, in our limited experience, appeared to be an attractive alternative: it is less invasive, with no morbidity and a shorter hospital stay.

Surgical correction of conotruncal anomalies with a malaligned ventricular septal defect involves formation of a tunnel to redirect the left ventricular blood through the septal defect to the aorta. Hence, the systemic outflow tract is formed mainly by a ventricular septal defect patch. A ventricular intramural septal defect results when the patch is sutured to the free-wall trabeculations of the right ventricle near the aortic valve; consequently, blood can flow from the new left ventricular outflow tract, between the trabeculae, into the right ventricle.[1] It has been suggested that suturing the patch to the ventriculo-infundibular fold, as close as possible to the aortic valve and not to the trabeculae of the free right ventricle wall, should prevent ventricular intramural septal defect formation.[2]

A residual intramural ventricular septal defect should be suspected in the presence of septal defect shunt outside the plane of the intraventricular septum after biventricular repair of a conotruncal anomaly. These defects are difficult to localise and may lead to multiple re-operations, with high risks for morbidity and mortality.[1] In our experience, the first clue to the existence of an intramural ventricular septal defect is the presence of red matter in the right ventricle at systole on the apical four-chamber view in a transthoracic echocardiography, which indicates the presence of a septal defect above the surgical patch outside the plane of the intraventricular septum. This finding led us to examine the tract of the shunt through the free right ventricular wall above the ventricular septal defect patch in the sub-costal coronal five-chamber view. While the shunt in post-surgical defects usually tracts around the patch, it can tract abnormally in the free right ventricular wall above the defect patch. This must be differentiated from normal tricuspid valve diastolic inflow.

Transoesophageal echocardiography is usually sensitive for detecting intraoperative and early post-surgical residual ventricular septal defects,[8] but the defect may not appear initially because of right ventricular hypertrophy. Only with regression of septal hypertrophy does an intramural ventricular septal defect shunt begin to tract through the free right ventricular wall, and this can easily be detected by transthoracic echocardiography during follow-up. Surgical correction of an intramural ventricular septal defect involves detaching the previously positioned patch and reattaching it to the ventriculo-infundibular fold proximal to the trabeculae.[2]

Conclusions

Residual intramural ventricular septal defect is a rare finding after repair of conotruncal anomalies. It should be suspected when the residual ventricular septal defect lies outside the plane of the interventricular septum. Surgical repair of such defects is effective but is associated with high risks for morbidity and mortality, and the transcatheter approach may offer an attractive alternative. The size of the defect may predict which patients are symptomatic. In our limited experience, a patient with a ventricular septal defect of more than 4 mm is likely to present later with symptoms of congestive heart failure. We recommend that ventricular septal defect patch closure be conducted if intra-operative transoesophageal echocardiography shows a ventricular septal defect shunt of 4 mm or more. A study with a larger number of patients is needed to confirm this conclusion.

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